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One-stage surgical removal of post-hysterectomy intravenous leiomyomatosis with inferior vena cava and heart extension using normothermic cardiopulmonary bypass: A case report

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Abstract

Intravenous leiomyomatosis (IVL) is an extremely rare pathological condition with undetermined ethiopathogenesis. Successful management relies on complete surgical tumor excision, which can be performed as a one or two-staged procedure. This report presents a case of 52-year old woman with diffuse IVL extending to the right cardiac cavity. The patient underwent a combined single stage procedure by establishing normothermic cardiopulmonary bypass with tumor excision through a right retroperitoneal approach. She continues to remain well 6 months following her hospital discharge with imaging revealed complete tumor resection. Our report demonstrated efficient one-stage surgical removal of intracardiac leiomyomatosis using normothermic cardiopulmonary bypass. Clinicians must be aware of this condition when patient have a history of uterine fibroids or uterine surgery, accompanied by deep vein thrombosis, tumor in right cardiac cavity or right heart dysfunction.

Key words

intravenous leiomyomatosis, intracardiac extension, normothermic cardiopulmonary bypass, uterine fibroids

Introduction

Intravenous leiomyomatosis (IVL) is an extremely rare pathological condition originating from myometrial veins and composed of smooth muscle cells (1). Although IVL is histologically benign, the nodular masses may extend into the inferior vena cava (IVC), subclavian vein, right cardiac cavity or lungs (2-4). Intracardiac extension of IVL was first described by Birch at the beginning of the 20th century (5) and less than 100 cases have been reported in the literature to date (6). The patient presentation may vary widely, aggravating an early diagnostic process because patients may be asymptomatic despite extensive intravenous extension. Intracardiac involvement may result with sudden fatal outcome due to right-sided congestive heart failure, emphasizing importance of prompt and accurate disease recognition (7).

The etiopathogenesis of IVL has yet to be determined, however, in the majority of cases the patients have a history of uterine fibroids or disease (8).

Successful management relies on complete surgical tumor excision, which can be performed as a one or two-staged procedure, involving resection of the pelvic tumor and upward dislodgment of the intracardiac and intravascular tumor by opening the right atrium (9,10).

The aim of this report is to present a case of diffuse IVL extending to the right cardiac cavity, which was successfully resected using normothermic cardiopulmonary bypass with induced left ventricular (LV) fibrillation. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Case report

A 52-year old female patient (gravida 3, para 2) was referred to our Department with the mass in the right atrium (RA) arising from inferior vena cava (IVC), presented without any signs nor symptoms.

Her past surgical history revealed total abdominal hysterectomy (TAH) for leiomyoma 12 years before admission. Furthermore, her past medical history was significant for three-year hematologist follow up due to the suspicion of the right ovarian vein and IVC thrombosis. She was on chronic anticoagulation therapy and had no other history of major systemic disease and cardiovascular risk factors, respectively.

Computed tomography angiography (CTA) revealed 6 cm large retroperitoneal tumor next to right adnexa and 11 x 2 cm tumor in the IVC which invaded the RA. Magnetic resonance imaging (MRI) findings were consistent with CTA (Fig. 1a, 1b). A transthoracic echocardiography showed solid mass (2 x 1 cm) in the RA arising from the IVC.

After multidisciplinary consult of cardiac, vascular and gynecologist surgeons, the patient underwent prompt surgical procedure.

A median laparotomy was performed, as we opened retroperitoneal space and visualized myomatous formation around 6 cm invading right ovarian vein. After colon mobilization, IVC and aorta in level of right renal vein were visualized. Careful surgical dissection and resection of right ovarian vein was carried out. Subsequently, median sternotomy was performed and after heparinization, normothermic cardiopulmonary bypass (CPB) was established between the ascending aorta, superior vena cava (SVC) and IVC at the point just above the iliac vein confluence. After snaring of the SVC cannula, LV fibrillation was induced and the right atriotomy was performed to prevent embolization of the tumor mass followed by subsequent placement of the vascular clamp on the IVC ensuring total CPB.

Then we performed 4 cm long incision in the IVC extending just below right ovarian vein directed cranially and placed vascular clamp on the left renal vein to mitigate bloodless surgical field. The smooth, firm white-colored tumor mass was gently grasped with resano forceps and completely extracted from within the IVC in one piece under direct vision from the RA (Fig. 2). The length of the tumor from IVC to the RA was around 40 cm. We closed the RA and IVC in the running fashion and the patient was weaned from CPB without any difficulties. CPB and LV fibrillation times were 36 and 12 minutes, respectively. The total operative time was 105 minutes, and intraoperatively patient received 250 mL of red blood cells.

The postoperative period was unremarkable and the patient was discharged from hospital on the twelfth postoperative day.

She was maintained on regular anticoagulant warfarin treatment to achieve an international normalized ratio of 2 to 3. Postoperative CTA revealed complete tumor resection 6 months after procedure.

Discussion

One of the largest series of cases, published by Du and associates, reported incidence of IVL in less than 0.1% of excised leiomyomas (11). It preferentially involves premenopausal Caucasian women, usually reporting a past history of uterine fibroids, adenomyosis and hysterectomy. Mizuno and associates reported that more than a half of the patients with IVL have a prior history of hysterectomy (8).

Intracardiac extension of the IVL is encountered in approximately 10% of cases with the peak occurrence at the fifth decade in most studies (1,6,8,11). Lam et al. reported that IVL can

follow two well defined pathways into the systemic venous circulation: predominantly the uterine vein, and rarely the ovarian vein bypassing the iliac veins (1).

Although initially misdiagnosed and treated as an IVC thrombosis, disease progression to right heart chamber was asymptomatic in our report. Possible explanation was identified on preoperative MRI images which shown significantly enlarged azygos venous system. It represents an accessory venous pathway supplying an important collateral circulation between the superior and inferior vena cava (12). This theory also has a therapeutic implication: it allows surgeon to ligate infrarenal IVC without compromising the venous return. Furthermore, a nodular, solid mass in the right atrium occurred during transthoracic echocardiography was not large enough to fill the cardiac cavity and it was not attached to the endocardium. Based on our findings and other published literature, differential diagnosis is broad and includes common medical conditions (e.g. deep venous thrombosis) and unusual growth patterns such as diffuse peritoneal leiomyomatosis and benign metastasizing leiomyomas (6). IVL should also be considered in the differential diagnosis for nonthrombotic pulmonary embolism (13).

In the present case, we apply one-stage surgical approach; the pelvic and chest surgeries were performed at the same time. The first successful operative resection of IVL with cardiac extension was performed as a multistage operative procedure, back in 1980 (14). Over the years, surgical strategies in this particular setting have evolved. In the past decade, a two-staged procedure was more preferable and appropriate in patients with poor general condition, large vascular tumor, pelvic adhesions and hemodynamic instability (7). First stage operation usually involved atriotomy and in a 6-week time span patient received antiestrogen therapy to control residual vascular tumor. In rare cases it may even result with tumor regression (7). One-stage surgical procedure offers potential benefits over two-staged such as reduction of intraoperative thrombosis events with similar intraoperative and postoperative complication

rates (7,11). Procedures that do not require CPB have been reported, chiefly when the tumor is of small diameter (10).

Whether one or two-staged approach is selected, exact recurrence rate of completely resected IVL is still unknown. Current literature findings suggest possibility of micro metastatic disease in the lumen of veins which offers potential for tumor regrowth (1,11). However, up to 30% of patients with incomplete tumor resection experience recurrence, emphasizing importance of adjuvant hormonal therapy (6,7). Du and associates report recurrence rate of 16.6% in their large case series study, implicating young age and size of the initial tumor as a predisposing factor (11). Although adjuvant hormonal therapy has been used in several different forms, evidence regarding their efficacy is inconsistent (7).

A preoperative clinical and imaging assessment made our decision towards one-stage approach, using normothermic CPB with LV fibrillation. This provides a bloodless field and excellent visualization for removal of the intravascular and the right atrium tumor. Furthermore, normothermic CPB with LV fibrillation help to avoid complications such as organ dysfunction and coagulopathy that can occur with deep hypothermic circulatory arrest (15).

Our report demonstrated efficient one-stage surgical removal of intracardiac leiomyomatosis using normothermic cardiopulmonary bypass. Clinicians must be aware of this condition when patient have a history of uterine fibroids or uterine surgery, accompanied by deep vein thrombosis, tumor in right cardiac cavity or right heart dysfunction. Prompt and accurate diagnosis to accomplish an exact treatment and later surveillance to ensure a minimal risk of recurrence is critical.

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We obtained a signed consent form from our presented patient.

Author contributions

Conception and design: GV, AŠM, MM, AZK.

Acquisition of data: GV, MM, ŽĐ.

Analysis and interpretation of data: GV, IP, AZK.

Drafting of the manuscript: MM, ŽĐ.

Critical revision of the manuscript for important intellectual content: GV, AŠM, IP, AZK.

Supervision: GV, AZK.

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Figure legends

Figure 1a. Preoperative coronal plane magnetic resonance (MRI) demonstrates a mass present in the right ovarian vein, as well as within the inferior vena cava.

Figure 1b. Intracardiac tumor extension from the inferior vena cava.

Figure 2. Right ovarian vein leiomyoma and intravenous leiomyomatosis.